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Not Just Kids –A Case of Adult IgA Vasculitis

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INTRODUCTION

IgA vasculitis, formerly known as Henloch Schonlein Purpura, is generally recognized as the most common form of systemic vasculitis in children, with 90% of cases occurring in the pediatric age group. However, the remaining 10% are noted in adult populations, and often has poorer long term outcomes if not identified and treated quickly. IgAV is commonly associated with streptococcal infections, although additional triggers such as drug ingestions, insect bites, and vaccinations have been described. Symptoms can include palpable purpura, acute abdominal pain, arthralgias, and hematuria.

Recognizing the constellation of signs and symptoms is key for expediting appropriate therapy, which may reduce long term renal damage.

CASE REPORT

A 27-year-old man with a history of asthma presented to the ED for maculopapular rash, polyarthralgias, and myopathy.

- Found to be hypertensive (160s/100s) and tachycardic (115)
- Creatinine 1.3, ESR 13, and CRP >80
- History was significant for an upper respiratory illness 10 days prior, camping trip 5 days prior with ingestion of several drugs including cocaine, MDMA, marijuana and psychogenic mushrooms.

He was admitted overnight for rheumatologic and infectious disease workup, but discharged the following day with plan for outpatient follow up.

Two days later, he again presented to the ED with acute abdominal pain and nausea with multiple episodes of vomiting, and was readmitted.

- Persistently hypertensive (160s/100s)
- Worsened purpuric rash
- Nonpitting edema in upper and lower extremities
- Creatinine 1.2, normal CBC, ASO titer elevated (957)
- CT abdomen showed small bowel edema, and EGD/colonoscopy were inconclusive

His collective symptoms were reviewed and included the following:

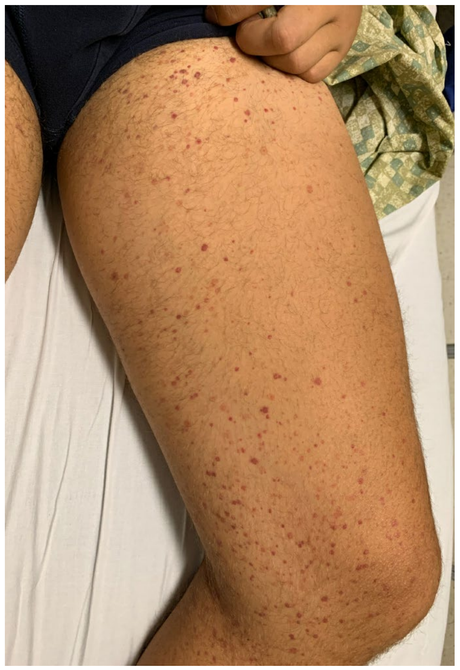
- Palpable purpura without thrombocytopenia or coagulopathy
- Arthralgias
- Abdominal pain

IgAV was hypothesized and skin/renal biopsies were obtained.

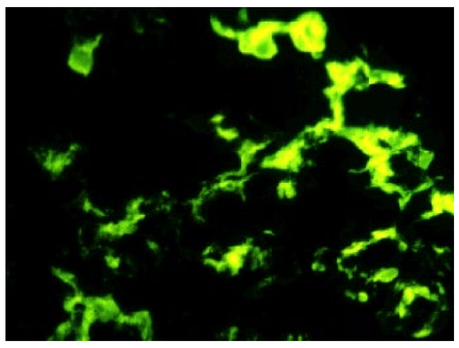
- Skin biopsy: "leukocytoclastic vasculitis with granular IgA deposition in superficial vascular walls"
- Renal biopsy: "positive mesangial immunofluorescence for IgA"

Based on these findings in conjunction with his symptoms and lab findings, he was started on a long steroid taper with plan for close outpatient follow-up.

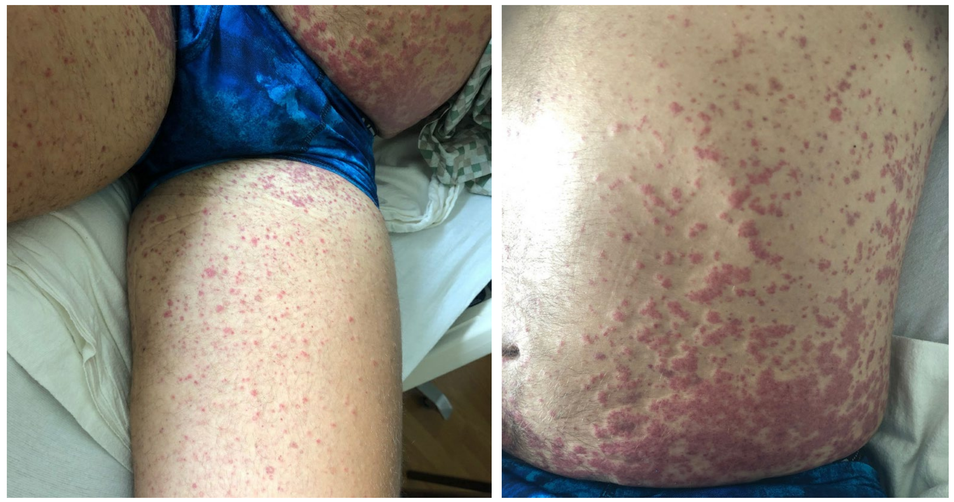
IMAGES



Hospital Day 1



Mesangial IgA immunofluorescence



Hospital Day 3

HOW TO FOLLOW UP:

- ❑ Monitor closely for signs of renal disease progression:
  - ❖ Elevated serum creatinine concentration (>1.25)
  - ❖ Hypertension (>140/90 mmHg)
  - ❖ Persistent (eg, for more than six months) protein excretion above 1000 mg/day
- ❑ Manage hypertension aggressively with ACEI (and additional agents if needed) to minimize additional renal damage
- ❑ If patients have persistent overt proteinuria and/or an elevated serum creatinine concentration, consider referral to nephrology. Progression to end-stage renal disease is approximately 15–25% at 10 years and 20-30% at 20 years.

DISCUSSION

IgA vasculitis can affect many organ systems including integumentary, GI, and renal. Although it is most common in children age 5-9, it can affect adults and requires prompt recognition to facilitate appropriate therapy.

The classic tetrad of symptoms includes:

- Palpable purpura without thrombocytopenia and/or coagulopathy
- Arthralgias
- Abdominal pain
- Renal disease

These symptoms often present in a predictable order, with purpura noted 4 days prior to other symptoms.

Our patient required careful blood pressure management due to his renal involvement (started on ACEI), and steroids were initiated. At time of discharge, his renal function and rash were improving, and abdominal pain had resolved. Although full renal recovery is common in up to 89% of patients, long term renal impairment may occur. Close monitoring is imperative to minimize further renal damage.